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CASE REPORT

The EXIT (*Ex Utero* Intrapartum Treatment) Procedure

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Fetuses with upper airway obstruction have a high mortality rate if proper airway management is not immediately administered after delivery. The EXIT (*ex utero* intrapartum treatment) procedure is a new technique that establishes the fetal airway while uteroplacental circulation is still maintained. The prognosis of such neonates has much improved after the introduction of this procedure. We report two neonates with prenatal diagnosis of upper airway obstruction; they were born smoothly via the EXIT procedure. The first was a case of epignathus, a rare type of nasopharyngeal or oropharyngeal teratoma derived from all three germ cell layers with variable maturity. The second was a case of giant lymphangioma that resulted from localized malformations in the development of the lymphatic system. Furthermore, compared with routine cesarean section, the short-term maternal outcomes were not different with regard to infection rate, estimated blood loss, wound complication, and postpartum hospital stay. [*J Formos Med Assoc* 2008;107(9): 745–748]

Key Words: epignathus, EXIT procedure, lymphangioma

Just born neonates with upper airway obstruction are at high risk of neonatal mortality and morbidity because of perinatal hypoxia; therefore, immediate airway intervention is required. Recent advances in prenatal sonography and magnetic resonance imaging (MRI) have improved the prenatal diagnosis of such congenital abnormalities, thus improving neonatal outcome after a complete peripartum management plan.

The EXIT (*ex utero* intrapartum treatment) procedure is a successful technique for establishing patent airway in infants with obstructive airway when placental circulation is still being supported. It was first performed by Norris et al in 1989 for a case of cervical teratoma.[1] Later, in 1997, Mychaliska et al standardized the procedure and provided guidelines.[2] This approach requires multidisciplinary cooperation among the obstetrician, neonatologist, anesthesiologist, and otolaryngologist.

Case Reports

Case 1 (Figure 1A) was a female neonate born at 36 weeks' gestation, with a body weight at birth of 1780 g, to a 35-year-old healthy mother. One oral cavity mass had been noted on routine prenatal ultrasound examination at 30 weeks of gestation. The mass lesion had enlarged progressively, causing polyhydramnios and intrauterine growth retardation. Pelvic MRI showed a heterogeneous mass with cyst components protruding from the oral cavity. Epignathus with upper airway obstruction

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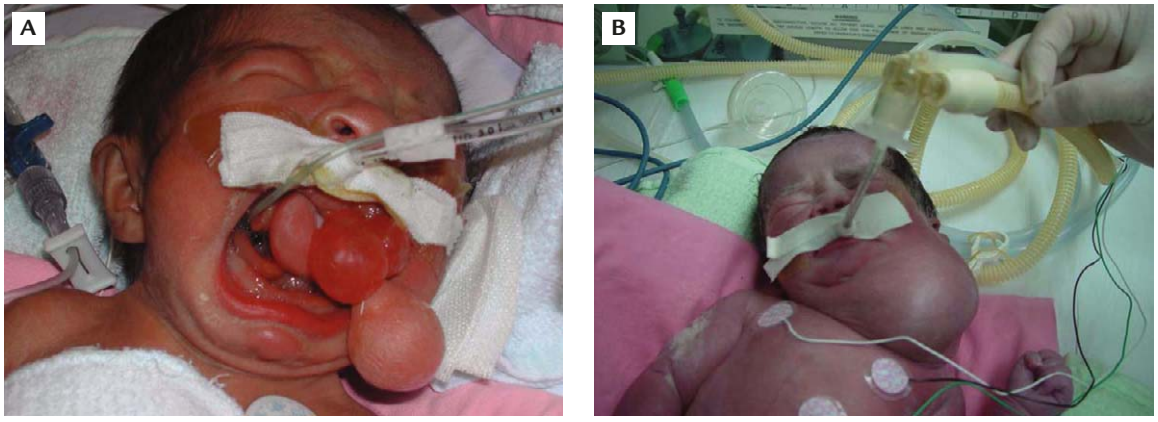

Figure 1. (A) Case 1: after delivery via the EXIT procedure, an oral tumor with different components (a 2 × 2 cm solid mass with skin appendage and peduncle, 2 cyst-like masses each measuring 1 × 1 cm) was noted, which arose from the hard palate with connection to the mouth floor. (B) Case 2: after delivery via the EXIT procedure, a huge and soft mass with smooth surface was noted at the left neck, measuring 10 × 5 × 4 cm with a 6 × 4 cm tense portion.

was suspected. After discussion with otolaryngologists and neonatologist, an EXIT procedure was planned.

Case 2 (Figure 1B) was a female neonate born at 36 weeks' gestation, with a body weight at birth of 3034 g, to a 32-year-old healthy mother. A left neck mass had been noted at 20 weeks' gestation on routine prenatal sonography. It had grown progressively in size. Maternal pelvic MRI showed an ill-marginated, lobulated, and septated mass lesion at the left neck with downward extension into the left mediastinum. A huge lymphangioma with upper airway compression was impressed and an EXIT procedure was scheduled.

Both cases were delivered via cesarean section 20–30 minutes after the mothers were under deep general anesthesia. As the head, chest and right arm of each neonate was being delivered, the umbilical cord was not clamped to maintain adequate maternal–fetal circulation and fetal oxygenation. Meanwhile, a pulse oximeter was applied on the baby's right forearm aseptically to monitor fetal heart beat and oxygen saturation. Extrauterine intrapartum intubation was performed smoothly after bronchoscope examination. After establishing the airway, the umbilical cord was interrupted and the baby was completely delivered.

The postnatal conditions of the two neonates were stationary. They received tumor excision later after completion of evaluation. The final pathology reports were dermoid cyst, one subtype of teratoma, and cystic lymphangioma, respectively. In the first neonate, other multiple congenital anomalies including ventricular septal defect, enlarged third ventricle of the brain and hiatal hernia with severe gastroesophageal reflux were also documented. Unfortunately, the first infant died at the age of 8 months due to repeated aspiration pneumonia with sepsis. The second infant died at the age of 3 months because of intractable lymphangioma with tracheobronchomalasia, even after repeated local tapping, minocycline and OK432 sclerotherapy.

Discussion

The EXIT procedure is used in neonates with upper airway obstruction and improves the infants' outcomes significantly.[3–5] The indications have been expanded successfully to include fetal neck mass, congenital high airway obstruction syndrome (CHAOS), reversal of tracheal occlusion for congenital diaphragmatic hernia, unilateral pulmonary agenesis, and EXIT-to-extracorporeal membrane oxygenation.[3,4,6] This procedure requires expertise and careful preparation. Figure 2 illustrates the organization of specialists involved in the operation.

The EXIT procedure is quite different from cesarean section in several ways and requires close monitoring. Tables 1 and 2 summarize the fetal

and maternal vital signs and morbidity during and after the EXIT procedure. With regard to the parturient, deep anesthesia is important to enhance uterine relaxation and preserve uteroplacental circulation.[5,7] However, maternal hypotension and uterine atony causing massive bleeding are major problems. According to the suggestion of Midrio et al,[7] continuous ephedrine infusion was administered to the first mother to keep systolic blood pressure between 120 and 140 mmHg. Once the infant was delivered completely, pitocin was administered to the mother before clamping of the umbilical cord to prevent uterine atony and minimize maternal blood loss.[8]

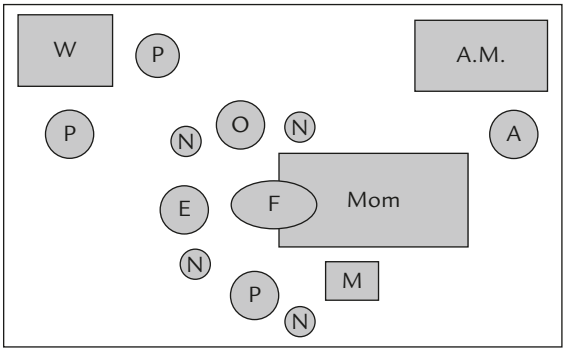

Figure 2. Organization of the EXIT team in the operating room. W = warmer; P = pediatrician or neonatologist; N = nurse; O = obstetrician; E = otolaryngologist; M = monitor; A = anesthesiologist; A.M. = anesthetic machine; Mom = mother; F = fetus.

The average estimated maternal blood loss for those undergoing the EXIT procedure is about 848.3 ± 574 mL to 970 ± 510 mL, and the average time on placenta bypass is 30.7 ± 17.8 minutes to 45 ± 25 minutes.[3,6,9] In our cases, total maternal blood loss was estimated to be 800 mL in the first case and 400 mL in the second case (Table 2). The duration from uterine incision to umbilical cord clamping was 15 minutes and 6 minutes, respectively (Table 1).

With regard to the fetus, maternal deep anesthesia is also important to reduce fetal spontaneous movement for rapid sequence intubation.

Table 1. Basic characteristics and hemodynamics of the neonates during the EXIT procedure

	First neonate	Second neonate
Saturation (%)	60–76	80–88
Heart rate (beats/min)	140–160	140–160
Gestational age (wk)	36	36
Birth body weight (g)	1780	3034
Duration on placental support (min)	15	6
Apgar score (1 min → 5 min)	6 → 7	6 → 8
Prenatal complications	Polyhydramnios	–

EXIT = ex utero intrapartum treatment.

Table 2. Maternal vital signs and hemogram during the EXIT procedure

	First mother	Second mother
Heart rate (beats/min)	80–95	80–100
Systolic blood pressure (mmHg)	120–140	125–140
Body temperature (°C)	35.7–35.8	35.6–35.8
Estimated blood loss (mL)	800	400
Length of operation (min)	105	75
Hemogram (mg/dL) (pre-op → post-op)	10.3 → 6.0	10.7 → 9.8
Hematocrit (%) (pre-op → post-op)	29.9 → 18.2	32.8 → 29.9
Blood transfusion	–	–
Wound complication	–	–
Postpartum hospital stay (d)	5	5

EXIT = ex utero intrapartum treatment.

Hence, the newborn's Apgar score will be underestimated.[5] In our cases, the 1- and 5-minute Apgar scores after complete delivery were 6 and 7 for the first baby and 6 and 8 for the second baby (Table 1).

Fetal pulsation and arterial saturation are monitored continuously by using a pulse oximeter. Aseptic procedure is important to reduce infection rate. We used a sterile protective plastic sleeve to cover the lead of the pulse oximeter, and then applied it to the exposed fetal hand. During the entire procedure, fetal arterial saturation was around 60–88%, and mean fetal heart rate was around 140–160 beats per minute (Table 1), which are compatible with other reports.[3,10] Because of high potential for poor peripheral perfusion, which would interfere with saturation measurement, Tanaka et al[11] suggested monitoring fetal hemodynamics with a portable Doppler and measuring cord blood gas. However, we did not adopt this method because it may increase the rate of infection and prolong the delivery course.

To prevent fetal hypothermia during the EXIT procedure, the operation room temperature should be kept at around 29.5–32.2°C. Continuous uterus irrigation with warm normal saline or Ringers' lactate solution and covering the exposed fetal portion with a sterile warm towel are suggested.[3–5] In addition, amnioreduction was done during partum for the first case because of severe polyhydramnios to reduce placental compression.[5]

Some studies report that the short-term maternal outcomes with EXIT are not different from those after routine cesarean delivery with regard to infection rate, estimated blood loss, wound complication, and postpartum hospital stay.[3,9] However, Hirose et al[6] reported that wound complications and estimated blood loss were higher in the EXIT group. In our first case, maternal hemoglobin decreased from 10.3 mg/dL to 6.0 mg/dL. However, the postpartum vital signs of this mother were stable; she did not have dizziness, hypotension, or tachycardia under supportive treatment. Hence, blood transfusion was not given.

In conclusion, short-term fetal outcomes are good after successfully establishing an airway and the maternal risks are decreased under a well-planned and organized EXIT procedure. However, the medium and long-term fetal outcomes are still strongly related to the underlying disease or other associated congenital anomalies.

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